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IMAGES IN CARDIOLOGY

Noncompaction Cardiomyopathy and Multiple Coronary Arterioventricular Fistulae

1 or 2 Distinct Disease Entities?

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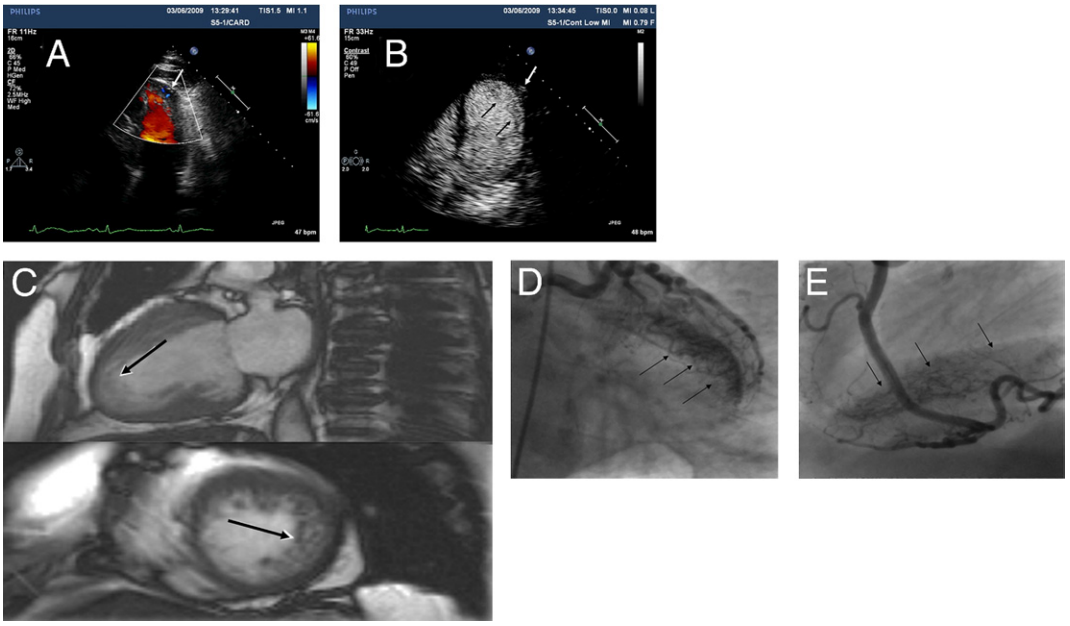

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A 67-year-old man presented to our department with progressing dyspnea and effort angina for the past 3 months. The transthoracic echocardiogram showed hypertrabeculation of the left ventricular lateroapical region (**A and B**; Online Videos 1, 2, 3, 4, and 5) with a maximal ratio of noncompacted-to-compacted myocardium of 2.3. On cardiovascular magnetic resonance imaging **(C),** the ratio was 2.6, supporting the diagnosis of noncompaction cardiomyopathy. Both imaging modalities clearly depicted the endocardial border of the noncompacted myocardium **(arrows)** and the flow within it. Interestingly, on coronary angiography, left ventricular opacification occurred after left and right coronary injections (**D and E, arrows**, Online Videos 6 and 7), disclosing multiple coronary-left ventricular fistulae. There was no epicardial coronary obstructive disease. Reviewing the color Doppler and contrast echocardiogram images, this coronary malformation could already have been suspected because an unusually evident diastolic flow within the compacted layer of the myocardium **(white arrows)** was clearly shown. To our knowledge, this is the first report describing the association of noncompaction cardiomyopathy and multiple coronary-left ventricular fistulae in the same patient. We think that this case could demonstrate an arrest in embryogenic development somewhere between the regression of the myocardial sinusoids and the compaction of the myocardium and may be a continuum of only 1 disease.